248 Liver function and vitamin D in children with cystic fibrosis

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Introduction: The liver is important in the metabolism of vitamin D especially its hydroxylation to 25 OH D3. Liver disease occurs in up to 15–20% of children with CF raising the possibility that Vitamin D levels may be influenced by their liver function. Our aim was to look at vitamin D levels in children and to correlate it with their liver function tests.
Methods: A retrospective review of Vitamin D levels and liver function tests (AST, ALT, GGT, Alk Phos, Protein and Albumin) was conducted. Seventy-seven children with mean age of 12 were included in study. Minitab Statistical Package was used to analyse the data. Regression analysis and independent t tests were carried out to determine any relationship between vitamin D and liver function.
Results: Eleven children (14%) had a diagnosis of CF related liver disease (CFLD). The mean vitamin D was 73.12 (±28 SD) with no difference in mean vitamin D between CFLD patients and none CFLD patients (mean of 65 nmol/ L versus 75 nmol/ L respectively, $p = 0.27$). Vitamin D negatively correlated with GGT, protein and ALT ($p = 0.02$, 0.04 and 0.02 respectively). Vitamin D did not correlate with albumin, AST or Alk Phos ($p = 0.93$, 0.68 and 0.56 respectively). When those with CFLD were removed from the data, vitamin D still negatively correlated with protein ($p = 0.04$) and GGT ($p = 0.03$).
Conclusion: The cause of low vitamin D in CF is multifactorial. This study provides some evidence of a direct relationship (either cause or effect) between vitamin D and liver function. Further research is needed to determine if increased vitamin D supplementation in CF would affect liver function.

249 Influence of calcium and vitamin D on bone mineral density in children with cystic fibrosis

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Low bone mineral density (BMD) is relatively frequent finding among patients with cystic fibrosis (CF). This complication is related with age, severity of lung damage and nutrition disorders.
Aim: To evaluate the effect of calcium and vitamin D supplementation on low BMD in patients with CF.
Materials and Methods: Z-score of BMD was assessed in 47 patients with CF and was performed with Quantitative Ultrasound. Chest deformation was found in 1/3 of schoolchildren with CF. Children with CF were divided into two groups: first group: 26 children younger than 12 years; second group: 21 children older than 12 years. All patients were pancreatic insufficient and received adequate basic therapy including multivitamins. Calcium and vitamin D supplements were given to correct the low values of BMD in children with CF for six months.
Results: Initial Z-score showed decreased values in both study groups: −0.14±1.17 SD in CF children <12 years and −1.29±0.9 SD in CF patients >12 years. 25OHD values in the younger group were 29.3±15.2 ng/ml and 18.07±8.3 ng/ml in older group. Calcium values were similar in the two groups (2.4±0.1; 2.37±0.2). After six months of treatment with calcium and vitamin D supplementation, the control densitometry showed significant increase in Z-score to 0.9±1.5 SD in children <12 years and to −0.9±1.1 SD in older CF patients.
Conclusion: Calcium and vitamin D have positive effect on mineral bone density in children with CF.

250 Vitamin D deficiency and thoracic kyphosis in adult cystic fibrosis patients

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Objectives: This study tested the hypothesis that there is an association between vitamin D deficiency and thoracic kyphosis in adults with Cystic Fibrosis (CF).
Methods: Subjects were recruited during annual review at the National Referral Centre for Adult Cystic Fibrosis. Each subject had thoracic curvature measured using a flexible ruler (previously validated against lateral x-ray), and had venous blood sampling to measure vitamin D level.
Results: 36 patients were included in the study, M:F ratio = 1.64:1.
5.5% of patients were in the severe lung disease category while 44% and 50% had moderate and mild disease respectively. 40.5% of patients were kyphotic (thoracic curvature greater than 40º). 13.8% of patients had optimal vitamin D levels (>75 nmol/L) and 2.7% were severely deficient with a measurement <25 nmol/L. A weak correlation of $r = 0.17$ was found between angle of kyphosis and vitamin D levels. 43% of those patients studied had exposure to oral steroids.
Conclusions: The prevalence of vitamin D deficiency in this population was very high. But only weak correlation was found between the angle of thoracic kyphosis and vitamin D level. Further investigation would be of benefit to look at bone density and nutritional status in this population.

251 Should %ideal body weight (%IBW) or body mass index percentile (BMIp) be used to assess growth in children with cystic fibrosis?

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Background: There is strong evidence that BMIp predicts nutritional failure more sensitively and accurately than %IBW, having a stronger correlation with %FEV1. This study aims to determine which growth measurement correlates most closely with %FEV1 in a UK population. Studies comparing both methods have concluded there is little difference for children of average height, but in short and tall children, underweight can be misclassified. %IBW is the main growth outcome measure in the 2002 CF Trust guidelines, which are currently under review.
Methods: Data was obtained from the 2010 South & South West regional CF database. Height, weight, BMIp and %FEV1 were calculated at annual review. BMIp was converted to Standard Deviation Scores (SDS) using UK reference data.
Results: Data was collected from 263 children (age range 5–17 years). Median %IBW was 101 (SD ±16.17, range 67–198). BMI SDS range was −3.21 to 3.57 (median −0.08±1.13). Median %FEV1 was 87 (SD ±19.8, range 28–129). Overall, both %IBW and BMI SDS had a significant association with %FEV1, with BMI SDS having a stronger correlation ($r = 0.3$, $p < 0.01$). In children between 5–12 years old ($n = 115$), the association between BMI SDS and %FEV1 was weaker ($r = 0.18$, $p < 0.05$), with no significant association between %IBW and %FEV1. In children with height >75th percentile ($n = 43$), neither growth measurement was significantly associated with %FEV1.
Conclusion: Compared to %IBW, BMIp is more closely associated with lung function in children <75th percentile for height. The lack of correlation with either growth measurement and %FEV1 in tall children raises the possibility that other growth measures should be employed in this group.